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Case Report

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Approach to anal sphincter dysfunctions in a patient with endorectal pull-through procedure

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ABSTRACT

A seven-year-old male patient was examined in our pediatric surgery outpatient unit with abdominal distension. He had been operated on for congenital megacolon in the newborn period in another center. After a period of follow-up, he had been re-operated because of a diagnosis of insufficient pull-through, and a second pull-through had been performed. Following this procedure, myectomy and temporary colostomy procedures were reported in a different hospital. After all these treatments, he still had a distension episode, even associated with enterocolitis. In our clinic, we medically treated enterocolitis four times in 5 months. According to the council decision in our department, we decided to use a medically used Botulinum toxin to evaluate the possibility of internal anal sphincter achalasia (IASA), and therefore we injected Botulinum toxin into the internal sphincter under anesthesia. Following the procedure, the patient was brought to our attention only once in 4 months with distension. This data encouraged us to diagnose the disease and planned to perform partial sphincterotomy and internal sphincter myectomy. Nine months after the myectomy, the patient was not hospitalized and had a mild distension once. There is a significant improvement in his life quality; as a clear result, we observed and experienced that sphincter-related problems must be taken into account in incurable postoperative obstructive situations related to Hirschsprung's disease.

Keywords: Hirschsprung's disease, internal anal sphincter, achalasia

INTRODUCTION

Persisting obstructive diseases may lead to fecal stasis, which may progress to cases of enterocolitis associated with Hirschsprung's disease.[1] It is known that 11-55% of enterocolitis attacks may occur after surgery, and obstructive problems may continue.[1-5] Many methods have been tried to treat this condition.[1] These include redoing pull-through metronidazole treatment, preventive rectal irrigations, and the use of probiotics.[1,2,5]

Another method described for treatment is *Botulinum* toxin injection (Botox), which Langer described in 1997.[1,3] *Botulinum* toxin injection can be applied if complaints of enterocolitis continue despite trying the other methods mentioned above.[2,4] However, there are doubts about *Botulinum* toxin use.[5] Due to the effectiveness of *Botulinum* toxin, in cases where the injection is beneficial, continued injections will be required to maintain this therapeutic effect.[2] In other words, it is not a sustainable practice for every patient at all times. Alternatives to Botox are the topical application of nitric oxide to the anal canal, posterior myotomy, redo pull-through, or diverting colostomy.[2]

Posterior myectomy has been a recognized practice, especially for internal anal sphincter achalasia (IASA), for over 50 years.[6] Hurst first described internal anal sphincter achalasia in 1934.[6] It is a problem with the relaxation mechanism of the sphincter. It has been detected in 2.9-4.5% of children with chronic constipation, even without any other disease.[6,7] It is stated that this amount is a level that cannot be underestimated.[7] It is stated that it is characterized by the absence of the rectoanal inhibition reflex (RAIR).[6]

In the end, whatever procedures are performed, continuity of IASAs is possible, although all alternative treatments are used.[6] Besides, these applications also have their characteristic complications.[2]

In our case, we use the *Botulinum* toxin as a diagnostic tool rather than a treatment instrument for IASA. The report aimed to share our experience of diagnosing IASA with *Botulinum* toxin, and we discussed our findings with literature.

CASE

A seven-year-old male patient was hospitalized with abdominal distension and inflammatory findings. A high level of C-RP was found at 91 mg/ml (n: 0.15-5), and leucocytosis 18.65 U/L (n: 4.6-10.2) was detected. A plain abdomen X-Ray showed distension and obstruction (**Figure**). Enterocolitis was diagnosed as an exploding stool with a nasty smell that was found in a rectal digital examination. Broad-spectrum antibiotics and other procedural treatments for inflammation were performed.

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Figure. Abdominal distension in plain abdomen X-Ray

The patient had surgical procedures for congenital megacolon disease in some other hospitals. Endorectal Pull-through had been performed twice in a center and myectomy was also done in the same hospital. Pathology specimen of excised bowel obtained in the second pull-through, which was done as an expectation of remnant for an aganglionic segment, had resulted as usual. Unfortunately, as complaints continued, a colostomy was performed in another hospital. After a year, colostomy was closed in that hospital. However, he again suffered from the same problem.

In our hospital, encouraged after the patient's history, we thought that continuing problems despite all these performed procedures had to be related to sphincter mechanisms. To test our hypothesis, based on the literature, we decided to use medically used *Botulinum* toxin. We did not evaluate anorectal function with anal manometry because, in our facility, there is any. Also, a financial status of the family is not suitable to have an anorectal manometry in a different city. As a result, they requested this procedure to ease this social problem. After Botox, distension and clinical findings altered, were not totally disposed of, but were significantly reduced. In four months, the patient had mild complaints without needing clinical hospitalization.

We found this result satisfying enough to perform posterior partial sphincter myectomy. Myectomy containing internal sphincter muscles was performed in lithotomy position to the posterior region between four and eight o'clock location. The patient has been in a healed situation nine months after the surgery. Parents called the outpatient clinic by phone to ask for slight suffering and for some other complaints that were not related to the disease. Since the hospital discharge, they did not need to bring the patient for further examination.

DISCUSSION

After Hirschsprung's disease surgeries, even if the surgery is successful, complaints related to obstructive attacks such as distension, excessive gassiness, vomiting, and cramps may be encountered.[1] Obstruction findings that develop after pull-through may be due to technical reasons (structure, folding of the pull-through segment, Soave cuff) or functional reasons (anus internal sphincter hypertonicity).[1] It has been shown that there is no relationship between obstructive findings after surgery and the timing of the surgery.[4] In other words, there is an impression that postoperative Hirschsprung-related enterocolitis[1] may develop slightly fewer amounts in patients operated on at older ages than in the neonatal period.[4]

Internal anal sphincter achalasia (IASA) is at least one of the causes of childhood constipation and fecal obstruction.[6] IASA can also be caused by any functional or physical problem.[6] Although the exact cause is unknown, it is thought that IASA is caused by deterioration in the innervation of the muscles, nitrergic nerve deficiency, cholinergic hyperplasia, and abnormal peptidergic innervation.[6] This pathological condition may be encountered during Hirschsprung's' disease, but similar findings may occur without Hirschsprung's' disease.[6] The difference between them is that symptoms occur later in patients with constipation than in Hirschsprung's' disease.[7] In contrast, some studies state that patients with IASA have fecal incontinence; others state that the inability to defecate is evident.[7]

If this condition is left untreated, it can cause problems including enterocolitis, systemic inflammation, and mortality.[1] Studies have shown that enterocolitis attacks are reduced by approximately 17-76% with Botox.[1,5,7] There is no consensus regarding the amount and location of the injection.[1,6] There are studies with amounts between 30-200 units/ml and studies performed between 4-8 regions.[1,4] However, it is stated that symptoms may return within 3-6 months.[1] As another approach, it has been said that *Botulinum* toxin, nitroglycerin ointment, or calcium channel blockers can be used after the surgical procedure until the surgical area heals.[4]

There is also literature expressing negative opinions about the application. A study determined that Botox in the first month postoperatively did not reduce the likelihood of developing enterocolitis in patients who underwent Swenson surgery.[4] In a study, a group of patients who underwent posterior internal anal sphincter myectomy was compared with a group of patients who received *Botulinum* toxin, and similar results were reported.[6] In some studies, it has been stated that there is no need to perform Botox, as it has been stated that the expected effect of disrupting the anal sphincter appears over the years and only weakly.[7] For this reason, as general literature information, it has been claimed that procedures such as sphincterotomy or myectomy have begun to be performed instead of *Botulinum* injection.[4]

Various complications may occur with Botox. These include temporary incontinence, pelvic floor muscle paresis, rectal prolapses, and perianal abscess.[1,2] In addition, despite *Botulinum* toxin, there may be no benefit, and problems that may lead to repeat surgery may continue.[2]

The patient applied to our clinic after all IASA-related procedures such as redo pull-through, diverting colostomy, and posterior myectomy had been performed. However, there was no history of using nitric oxide topical cream. We injected the sphincter with *Botulinum* toxin to see whether the problem was IASA. When the patient showed significant clinical improvement, we decided that IASA caused it. Since the economic situation of the family would not buy it for continuous use of *Botulinum* toxin, we decided to perform a sphincter myectomy. It was observed that the patient's clinical well-being continued during follow-up.

CONCLUSION

Although there is enough information in the literature about the use of *Botulinum* toxin, it is clear that there needs to be a standard regarding what situations, at what intervals, and in what dosages it should be used. In our clinical experience, we have come to the understanding of identifying the IASA problem, if any, and combining it with sphincter myectomy application rather than using it as a treatment protocol. Such a method of use has also been suggested in the literature and has clinical use. This method is realistic in decision-making, and I believe that such use of *Botulinum* toxin will contribute to the search for treatment protocols.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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